1. You are shown an extremity radiograph on a 4-month-old with irritability and altered mental status. What is the MOST LIKELY diagnosis?

A. Rickets
B. Congenital syphilis
C. Non-accidental trauma
D. Scurvy

Rationales:
A. Incorrect. Rickets is the result of deficiency of vitamin D, and in turn leads to failure of ossification of the physeal cartilage. This leads to loss of the radiodense zone of provisional calcification, and fraying of the metaphyses. Failure of mineralization leads to a widened appearance of the physis. These features are lacking in the test case.
B. Incorrect. Congenital syphilis is a transplacentally acquired infection, during the second or third trimester of pregnancy. A negative maternal syphilis test early in gestation therefore may not exclude the diagnosis in the infant, since maternal infection could have been acquired later in pregnancy. Skeletal manifestations typically develop in the infant at 6 – 8 weeks of age. Although syphilis may manifest s diffuse periostitis with periosteal reaction, the typical findings that are also present include radiolucent metaphyses, and destructive lesions in the medial tibial metaphysis, known as Wimberger sign. These typical findings of congenital syphilis are not present in the test case.
C. Correct. Correct answer as listed in the findings above.
D. Incorrect. Vitamin C deficiency, or scurvy, results in inability to synthesize normal collagen. The appearance of the bones is characterized by osteoporosis, with thin, sharp cortices. In the epiphyses, the outer border is sharply marginated, resembling a thin ring, the Ring of Wimberger. The zones of provisional calcification are preserved, but are underlined by a subjacent zone of mineralization, termed the scurvy line. Fragility of the tissues can result in extensive subperiosteal bleeding, and this can exhibit exuberant periosteal calcification during the healing phase. Although the periosteal reaction in the test case can resemble the calcification of subperiosteal hemorrhage in scurvy, this does not occur until the healing phase. Further, other findings of vitamin C deficiency—the lucent zone, Wimberger’s Ring, and diffuse osteoporosis—are absent.
2. Regarding congenital anomalies of the spinal cord, which of the following is CORRECT?

A. There is an increased incidence of tethering in patients with anal atresia.
B. Imaging of the cord is needed in infants with a low-lying coccygeal dimple.
C. Lipomyelomeningocele is associated with Arnold-Chiari malformation.
D. Skin-covered lesions are not associated with tethering.

Rationales:
A. **Correct.** Patients with anal atresia are at high risk of occult cord anomalies, resulting in tethering, and should undergo screening.
B. **Incorrect.** Low-lying dimples, unassociated with skin tags, skin discoloration, or hair tufts, are at low risk of tethering, and routine screening for these patients is not recommended.
C. **Incorrect.** Unlike the open defects, such as meningomyelocele, patients with lipomyelomeningocele do not typically have associated Chiari malformation.
D. **Incorrect.** Skin-covered cord lesions, including the lipomyeloecele and lipomyelomeningocele, result in cord tethering and symptoms resulting from cord ischemia due to stretching of the cord.

Citations:
3. Which statement is CORRECT concerning vascular rings?

A. A double aortic arch usually presents during puberty.
B. The double aortic arch encircles the trachea anterior to the esophagus.
C. Symptomatic pulmonary sling is associated with a right aortic arch.
D. A left aortic arch with an aberrant right subclavian artery is typically asymptomatic.

Rationales:
A. Incorrect. Double aortic arch is a true vascular ring resulting in compression of both trachea and esophagus. Symptoms often begin at birth and can include stridor, wheezing, vomiting, and dysphagia.
B. Incorrect. Double aortic arch typically encircles both the trachea anteriorly and esophagus posteriorly by its right and left arches. Lateral radiographs will demonstrate anterior bowing of the trachea. On barium swallow posterior indentation of the esophagus is present.
C. Incorrect. Anomalous origin of the left pulmonary artery is often an isolated finding though it may be part of a more complex anomaly. The left pulmonary artery arises from the right pulmonary artery crossing over the proximal portion of the right mainstem bronchus.
D. Correct. This most common congenital vascular anomaly is frequently found due to interruption of the right embryonic arch between the right common carotid and right subclavian arteries. The right ductus arteriosus usually involutes so no true vascular ring exists.

Citations:
Goo H, Perk T, Ok J et al. CT of congenital heart disease: Normal anatomy and typical pathologic conditions. Radiographics. 2003;23:S147-65
4. A neonate presents with complex congenital heart disease. Bilateral minor fissures are evident on chest radiographs. Which one of the following associated findings is MOST likely?

A. Biliary atresia
B. Malrotation
C. Interrupted inferior vena cava
D. Multiple spleens

Rationales:
A. Incorrect. Biliary atresia is seen in patients with polysplenia. This child has a heart lesion and trilobed lungs suggesting asplenia. Even in polysplenia, only a small fraction of children are affected. Thus, biliary atresia is not an expected abdominal finding in this child.

B. Correct. Although congenital heart disease is seen both in polysplenia and asplenia, the complex lesion in this child is more inconsistent with the severity of defects typically seen with asplenia than polysplenia. Children with polysplenia more often have simple shunts or anomalies of systemic or pulmonary return whereas those with asplenia often have transposition, common ventricles, and common atria. Bilateral minor fissures are indicative of bilateral trilobed lungs, which are consistent with asplenia syndrome (bilateral right-sidedness). Bilateral bilobed lungs are consistent with polysplenia syndrome (bilateral left-sidedness); however, often the fissures are difficult to see on neonatal films and thus lack of visualization of the minor fissure(s) does not equate to a diagnosis of polysplenia syndrome. Malrotation is present in most children with asplenia and thus would be an expected abdominal finding in this child. It should be noted that malrotation is also present in most children with polysplenia and thus is not a differentiating feature.

C. Incorrect. Interrupted inferior vena cava is a feature of polysplenia syndrome. This child has a heart lesion and trilobed lungs suggesting asplenia. In asplenia syndrome, the inferior vena cava is uninterrupted, although it is often malpositioned to the left rather than the normal location to the right of midline. Thus, an interrupted inferior vena cava is not an expected abdominal finding in this child.

D. Incorrect. Multiple spleens, of course, are consistent with polysplenia, not asplenia. Often the spleens are found in the right upper quadrant rather than the left, so absence of the spleen in its normal location is not sufficient evidence of asplenia. This child has a heart lesion and trilobed lungs suggesting asplenia, not polysplenia. Thus, multiple spleens are not an expected abdominal finding in this child.

Citations:
5. Regarding head sonography in the premature infant, which one of the following is CORRECT?

A. Periventricular leukomalacia manifests as cystic lesions.
B. Grade I germinal matrix hemorrhage is found at the occipital horns.
C. Lack of sulcation is suggestive of lissencephaly.
D. A cavum septum pellucidum suggests lobar holoprosencephaly.

Rationales:
A. Correct. Periventricular leukomalacia is an ischemic/hemorrhagic injury in the periventricular white matter, which evolves into multiple cystic spaces visible on sonography.
B. Incorrect. Grade I bleed is confined to the germinal matrix, and is typically found anterior to the foramina of Monro. By definition, blood does not extend into the ventricles.
C. Incorrect. Premature infants typically lack sulcation of the brain.
D. Incorrect. Cavum septum pellucidum and cavum vergae are normal findings at sonography in the premature infant.

Citations:

6. A newborn infant presents on the second day of life with a distended abdomen and bilious emesis. A radiograph of the abdomen demonstrates markedly distended intestinal loops throughout the abdomen. What is the BEST next diagnostic examination?

A. Upper GI study
B. Contrast enema
C. CT of the abdomen with IV contrast
D. Ultrasound examination of the abdomen

Rationales:
A. Incorrect. The case describes a distal bowel obstruction in a newborn. An upper GI study would not be warranted in cases of a newborn with a distal bowel obstruction.
B. Correct. The case describes a distal bowel obstruction in a newborn. The correct test of choice would be a contrast enema, preferably performed with water soluble, isotonic contrast in case of a perforation.
C. Incorrect. The case describes a distal bowel obstruction in a newborn. Abdominal CT plays no role in the workup.
D. Incorrect. The case describes a distal bowel obstruction in a newborn. Abdominal sonography plays no role in the workup, and the gas-filled bowel would render the examination difficult at best.
7. Concerning holoprosencephaly, which one is TRUE?

   A. The fornix and septum pellucidum are absent in all forms of holoprosencephaly.
   B. The posterior portion of the corpus callosum is absent in lobar holoprosencephaly.
   C. 10% of cases with holoprosencephaly are associated with cytogenetic abnormalities.
   D. Cebocephaly is most common in patients with lobar holoprosencephaly.

Rationales:
A. **Correct.** While the severity of holoprosencephaly varies from severe alobar to lobar, by definition, all forms of holoprosencephaly do not have a fornix or septum pellucidum, and may have hypoplasia of the pituitary and optic nerves.
B. **Incorrect.** The cerebral vesicles cleave in a dorsofrontal direction. In patients with lobar and semilobar holoprosencephaly, the posterior portion of the corpus callosum is present while the anterior portion is absent; the entire corpus callosum may be present in lobar holoprosencephaly.
C. **Incorrect.** Up to 40% of cases are linked to chromosomal anomalies, 20% with syndromes, with only 40% cases isolated.
D. **Incorrect.** Severe facial anomalies, such as cyclopia, cebocephaly and ethmocephaly, are most likely in patients with alobar holoprosencephaly. However, a normal face does not exclude a severe form of holoprosencephaly.

**Citations:**
Garel C, MRI of the fetal brain. Springer Verley New York 2004
8. Which one of the following findings is seen in most patients with Dandy Walker malformation?

A. Intact vermis
B. Small cisterna magna
C. Elevated tentorium cerebelli
D. Agenesis of the corpus callosum

Rationales:
A. Incorrect. By definition, the most characteristic finding of Dandy Walker malformation is complete or partial absence of the cerebellar vermis.
B. Incorrect. Fourth ventricle communicates with an enlarged cisterna magna bordered by the cerebellar hemispheres.
C. Correct. The tentorium and torcular are elevated with Dandy Walker malformation, best evaluated on midline sagittal images of the brain. On plain films, the anomaly is characterized by lambdoid-torcular inversion.
D. Incorrect. Dysgenesis of the corpus callosum occurs in only 20-25% of patients. Gyral abnormalities also occur in a small number of patients with dandy walker malformation.

Citations:
9. Which of the following radiographic findings is MOST useful in excluding the diagnosis of hyaline membrane disease in a newborn infant?

A. Pleural effusion
B. Hyperinflation
C. Unossified humeral epiphyses
D. Asymmetric pulmonary opacity

Rationales:
A. Incorrect. While pleural effusion is an uncommon finding in hyaline membrane disease, its presence or absence is not specific in excluding the diagnosis. Small pleural effusion may be seen in transient tachypnea of the newborn which can co-exist with hyaline membrane disease, especially on early radiographs. Although in the appropriate clinical setting, the presence of diffuse ground glass opacity with accompanying pleural effusion should raise the question of infection, particularly group B streptococcal pneumonia, pleural effusion alone cannot exclude hyaline membrane disease.
B. Correct. The underlying cause of hyaline membrane disease is deficiency of surfactant; this lipoprotein complex produced by Type II pneumocytes coats alveoli and prevents them from collapsing in expiration. Diffuse alveolar atelectasis with pulmonary underaeration is the hallmark of this disease. Increased lung volumes would strongly suggest excluding hyaline membrane disease as a diagnostic possibility. With early intubation and administration of exogenous surfactant, characteristic radiographic findings may be less marked and more transient.
C. Incorrect. Ossification of humeral epiphyses may be used as a sign of skeletal maturity on a chest radiograph. Ossified humeral epiphyses are not seen in infants less than thirty-six weeks of gestation and are present in approximately 40% of term infants. Therefore the presence of humeral epiphyses would diminish the likelihood of hyaline membrane disease, a disease of prematurity. Unossified epiphyses would not be useful in excluding the disease.
D. Incorrect. While diffuse symmetric ground glass opacity is the characteristic feature of hyaline membrane disease, some variation of this pattern is not uncommon. Lower lobe involvement tends to be more marked than upper lobe; opacity may be more marked on the right than on the left side. After surfactant administration there is often asymmetric clearing of the lungs. Therefore, the presence of asymmetry does not exclude the diagnosis of hyaline membrane disease.

Citations:
10. Concerning imaging of a child with a bronchial foreign body, which one is CORRECT?

A. A decubitus radiograph with the affected side up will be helpful in demonstrating ipsilateral air trapping.
B. A nuclear medicine ventilation perfusion scan will likely demonstrate symmetric lung perfusion but asymmetric ventilation.
C. On fluoroscopy the mediastinum will shift away from the affected side in expiration.
D. The majority of foreign bodies are radiopaque and visible on chest radiographs.

Rationales:
A. Incorrect. When placed in the decubitus position, the dependent (down) lung deaerates and the nondependent lung expands. Therefore to demonstrate air trapping i.e. lack of dependent deflation, the useful decubitus view is with the affected side down.
B. Incorrect. When the lung is abnormally ventilated there is also decreased perfusion due to reflex vasoconstriction. There may be very little perfusion of the affected side when significant air trapping is present.
C. Correct. This is one of the cardinal observations in fluoroscopic evaluation of a foreign body. On inspiration the mediastinum is in its normal location i.e. central. With expiration the mediastinum shifts towards the normal lung which deflates, and away from the persistently hyperinflated abnormal lung. Other fluoroscopic observations include lack of deflation and decreased diaphragmatic excursion on the affected side.
D. Incorrect. Only a small number of foreign bodies are radiopaque. The most common endobronchial foreign bodies are ingested foods, such as peanuts and raw carrots. Therefore the radiographic features rely on secondary findings, predominantly air trapping distal to the foreign body due to partial obstruction of the affected airway. With a larger or more long standing foreign body, pulmonary opacity due to atelectasis or post obstructive pneumonia may occur.

Citations: